A case of IgG 4 related disease

This 18-year-old female student with 1) IgG4-related disease with submandibular gland and parasellar tumor, complicated with right optic neuropathy, 2) atopic dermatitis was admitted via OPD due to scheduled rituximab therapy (C3D1). The patient had swelling neck masses over bilateral submandibular spaces 2 years ago. Moveover, right blurred vision was noted about 1 year ago. Then she went to our OPH OPD (2014/4/17) where Optic neuropathy of OD was impressed. Brain MRI revealed multilobulated tumors in the bilateral parasellar region, extension to right orbital apex, bilateral inferior orbital fissure and inferiorly to bilatearl petrygoid plate. Right optic nerve was encroached. Hence, she had admitted to NS ward (2014/6/15~2014/6/27) for right craniotomy with tumor removal 1.5 months ago. Pathology revealed small round blue cell tumor (SRBCT) composed of mainly polygonal to round tumor cells revealing scanty cytoplasm, high N/C ratio, r/o neuroblastoma, PNET, lymphoma, rhabdomyosarcoma or other undertermined malignancy. And the imunohistochemical stains also showed CD20 and CD3 (positive, patchy, but CD20>CD3), CD5(positive, focal), CD10(positive, focal), BCL2(negative), kappa and lambda light chain(equivocal, no definite light chain restriction), CD23(negative), while low-grade lymphoma cannot be ruled out. She was admitted to ENT ward (2014/7/14~2014/7/16) for incisional biopsy of right submandibular gland. The pathology report found abundant lymphoid cell and plasma cell infiltration in the submandibular gland with marked stromal fibrosclerosis change. The IHC stain revealed CD138(positive staining in the plasma cells), mixed CD3 & CD20-positive small lymphocytes, equivocal light chain restriction, IgG(positive, focal), and IgG4(positive, focally). Increased IgG4+ plasma cells (more than 50 IgG4+ cell/hpf) with IgG4+/IgG+ cell ratio > 40% are noted by immunohistochemical study, favoring IgG4-related sclerosing disease. Under the impression of IgG4-related disease, she was admitted and underwent Methylprednisolone 500mg pulse thrapy on 2014/8/13~15, cyclophosphamide pulse therapy (I; 500mg) on 2014/8/13, and Rituximab 500mg C1D1 on 2014/8/15 and C2D1 on 2015/5/11. Followed up brain MRI showed complete disappear of previously good enhancing soft tissue. Followed up visual field test also showed improvement of right visual field. This time, she was admitted for scheduled rituximab therapy (C3D1).